

1 **Supplemental Material**

2 **Table S1 Extrarenal manifestations based on ANCA serotypes in lupus nephritis**

	Total(N=95)	MPO-ANCA positivity(n=69)	PR3-ANCA positivity(n=26)	P Value
Fever(non-infectious)	24(25.3)	18(26.1)	6(23.1)	0.76
Malar rash	39(41.1)	28(40.6)	11(42.3)	0.88
Photosensitivity	7(7.4)	4(5.8)	3(11.5)	0.39
Mouth ulcer	11(11.6)	10(14.5)	1(3.8)	0.28
Alopecia	16(16.8)	13(18.8)	3(11.5)	0.54
Arthritis	49(51.6)	35(50.7)	14(53.8)	0.79
Serositis	16(16.8)	10(14.5)	6(23.1)	0.32
Neurological disorder	7(7.4)	5(7.2)	2(7.7)	0.90
Raynaud phenomenon	9(9.5)	8(11.6)	1(3.8)	0.44
Anemia	70(73.7)	49(71.0)	21(80.8)	0.44
Leukopenia	18(18.9)	12(17.4)	6(23.1)	0.53
Thrombocytopenia	24(25.3)	19(27.5)	5(19.2)	0.60

3 Note: Values are given as n (%).

4 ANCA, anti-neutrophil cytoplasmic antibody; MPO, myeloperoxidase; PR3, proteinase 3.

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1 **Table S2 Organs involvement potentially related to AAV based on ANCA serotypes in lupus nephritis**

	Total(N=95)	MPO-ANCA positivity(n=69)	PR3-ANCA positivity(n=26)	<i>P</i> Value
Skin lesions	5(5.3)	4(5.8)	1(3.8)	0.90
The eyes	0(0.0)	0(0.0)	0(0.0)	
Ear, nose, and throat	0(0.0)	0(0.0)	0(0.0)	
The pulmonary involvement	1(1.1)	1(1.4)	0(0.0)	0.90
Rapidly progressive glomerulonephritis	20(21.1)	14(20.3)	6(23.1)	0.77
The heart	0(0.0)	0(0.0)	0(0.0)	
Digestive system	0(0.0)	0(0.0)	0(0.0)	
Nervous system	0(0.0)	0(0.0)	0(0.0)	
The musculoskeletal	0(0.0)	0(0.0)	0(0.0)	
Other organs	0(0.0)	0(0.0)	0(0.0)	

2 Note: Values are given as n (%).

3 AAV, ANCA-associated vasculitis; ANCA, anti-neutrophil cytoplasmic antibody; MPO, myeloperoxidase; PR3, proteinase 3.

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1 **Table S3 Treatment regimens based on ANCA serotypes in lupus nephritis**

	Total(N=95)	MPO-ANCA positivity(n=69)	PR3-ANCA positivity(n=26)	P Value
Follow-up, mo	56.6(27.2 - 99.0)	62.1(26.0 - 147.0)	52.4(26.8 - 66.9)	0.11
Induction treatment				
Oral prednisone	95(100.0)	69(100.0)	26(100.0)	0.90
Cyclophosphamide	34(35.8)	30(43.5)	4(15.4)	0.02
Mycophenolate mofetil	34(35.8)	24(34.8)	10(38.5)	0.74
Tacrolimus	11(11.6)	8(11.6)	3(11.5)	0.90
Tripterygium wilfordii	28(29.5)	20(29.0)	8(30.8)	0.90
Maintenance treatment				
Oral prednisone	95(100.0)	69(100.0)	26(100.0)	0.90
Mycophenolate mofetil	27(28.4)	18(26.1)	9(34.6)	0.41
Tacrolimus	13(13.7)	10(14.5)	3(11.5)	0.90
Leflunomide	4(4.2)	2(2.9)	2(7.7)	0.30
Azathioprine	16(16.8)	14(20.3)	2(7.7)	0.22
Hydroxychloroquine	4(4.2)	3(4.3)	1(3.8)	0.90
Tripterygium wilfordii	48(50.5)	35(50.7)	13(50.0)	0.90

2 Note: Values are given as n (%) or median (interquartile range).

3 ANCA, anti-neutrophil cytoplasmic antibody; MPO, myeloperoxidase; PR3, proteinase 3.

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1 **Table S4 Univariate and Multivariate Cox regression of renal survival rates in ANCA-positive LN patients**

	Univariate analysis		Multivariate analysis	
	HR(95%CI)	<i>P</i> value	HR(95%CI)	<i>P</i> value
eGFR	0.97(0.94 - 0.99)	< 0.001	0.96 (0.94 – 0.98)	0.001
MPO-ANCA positivity	4.27(1.19 - 15.37)	0.03	-	-
CI ≥ 4 scores	6.24(2.12 - 18.38)	< 0.001	-	-

2 ANCA, anti-neutrophil cytoplasmic antibody; LN, lupus nephritis; HR, hazard ratio; 95% CI, 95% confidence interval; eGFR, estimated glomerular filtration rate; MPO,

3 myeloperoxidase; CI, chronicity index.

1 We compared PR3-ANCA-positive LN patients (n=26) with ANCA-negative LN patients
2 (n=1279), and provided more detailed about patient characteristics and pathological findings
3 (Table S5). Compared with ANCA-negative LN patients, PR3-ANCA-positive LN patients were older
4 (median: 36 [IQR, 24 – 42] versus 30 [IQR, 22 – 37] yrs; $P = 0.03$). Furthermore,
5 PR3-ANCA-positive LN patients had significantly lower serum concentrations of C3 and C4 at the
6 time of biopsy (median: 0.36 [IQR, 0.30 – 0.52] versus 0.47 [IQR, 0.36 – 0.68] g/L; $P = 0.01$) and
7 (median: 0.06 [IQR, 0.04 – 0.11] versus 0.11 [IQR, 0.06 – 0.17] g/L; $P = 0.001$). Conversely, there
8 was no significant difference in the sex, urine RBC count, proteinuria, SAlb, SCr, AI score, and CI
9 score. We analyzed renal outcomes between 26 PR3-ANCA-positive LN patients and 1279
10 ANCA-negative LN patients. There was no difference in the renal survival rates between the two
11 groups, as determined by Kaplan-Meier analysis. (Figure S1) There is some evidence to support a
12 different role for complement in AAV.^{7, 8} We need more investigations to explore the pathogenic
13 mechanism and renal outcomes about the different changes of complement in ANCA-positive LN
14 patients.

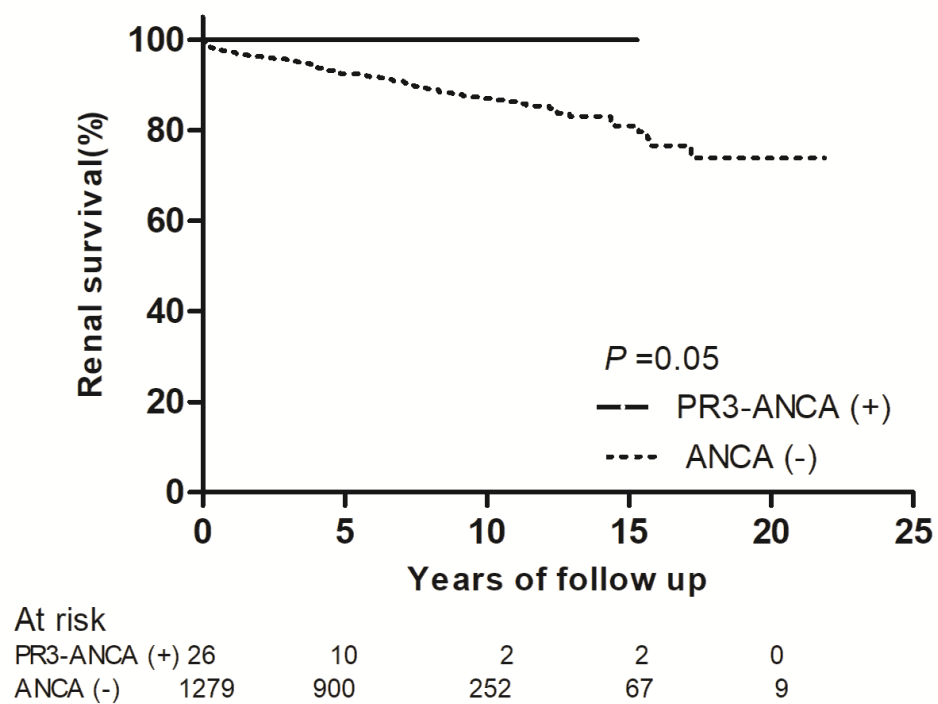
1 **Table S5 Comparison of clinic-pathological findings between PR3-ANCA positivity and ANCA**
2 **negativity in lupus patients**

	PR3-ANCA positivity (n=26)	ANCA negativity (n=1279)	P Value
<i>Patient characteristics</i>			
Men	1(3.8)	181(14.2)	0.16
Age, yrs	36(24 - 42)	30(22 - 37)	0.03
Proteinuria, g/24 h	2.3(1.5 - 4.5)	2.3(1.1 – 4.4)	0.70
Urine RBC count, ×10 ⁴ /mL	46(16 - 1869)	43(5 - 176)	0.06
SAlb, g/L	18.8(26.0- 32.8)	27.9(22.4 – 33.4)	0.17
SCr, μmol/L	74.3(60.6- 116.3)	73.4(54.8 – 101.7)	0.50
C3, g/L	0.36(0.30 - 0.52)	0.47(0.36 – 0.68)	0.01
C4, g/L	0.06(0.04 - 0.11)	0.11(0.06 – 0.17)	0.001
<i>Pathological findings</i>			
AI score	7(4 - 11)	7(2 - 11)	0.69
CI score	1(0 - 3)	2(0 - 3)	0.41

3 Note: Values are given as n (%) or median (interquartile range).

4 PR3, proteinase 3; ANCA, anti-neutrophil cytoplasmic antibody; RBC, red blood cell; SAlb, serum
5 albumin; SCr, serum creatinine; C3, complement 3; C4, complement 4; AI, activity index; CI,
6 chronicity index.

- 1 Figure S1 Comparison of renal survival rates between PR3-ANCA positivity and ANCA negativity
- 2 in lupus patients



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